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# A 68-YEAR OLD MAN WITH A CEREBELLOPONTINE ANGLE TUMOR

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## CLINICAL HISTORY

A 68-year-old diabetic male patient was initially admitted due to left-sided hearing loss, balance impairments and left-sided facial palsy in 2001. At that time he had been complaining of left-sided tinnitus for years. An MRI showed a mass in the cerebellopontine angle on the left side, causing mild compression of the brainstem. He underwent neurosurgery in 2002. The operation revealed that the 7th and 8th cranial nerves were profusely infiltrated by the tumor. The mass was excised by retrosigmoidal craniotomy and both nerves were preserved. Postoperatively, facial nerve palsy was still apparent. Gait and eyelid closure improved thereafter, whereas anacusis showed no improvement. He then underwent annual follow-up examinations. In 2006 T1-weighted horizontal MRI with gadolinium contrast enhancement demonstrated tumor recurrence. Moderate increase in growth was seen in 2007, but surgical intervention was not indicated at that time. In 2010 the patient suffered from increasing dizziness and balance impairment. MRI examination revealed tumor growth with medial expansion and brainstem compression (Fig. 1a) and second operation was performed. The tumor was again localized within the caudal cerebellopontine angle compressing the caudal brainstem and cranial nerves as well as the left crus cerebelli. On craniotomy the tumor was almost completely extirpated with a small residue remaining attached to the caudal cranial nerves. After operation, slight paresis of the 6th cranial nerve and hoarseness occurred.

## MICROSCOPIC PATHOLOGY

The formalin-fixed biopsy samples were processed with standard paraffin technique, and stained routinely (H&E, elastica van Gieson) as well as with antibodies against neurofilament, Ki67, vimentin, desmin, S-100β, smooth muscle actin, cytokeratins (clones MNF116, CK7), GFAP, CD34 and EMA. Both the initial as well as the recurrent tumor were composed of monomorphic spindle-shaped, often bipolar cells embedded within a myxoid stroma. Intermingled mature striated muscle fibers were seen, single or in small groups (Fig. 1b, 1c). The fine cytoplasmic tails were relatively short, and special architectures were not seen. Nuclei were round to oval and contained finely dispersed chromatin. Mitotic activity was absent. Blood vessels showed fibrosis. Lymphocytic infiltrates were apparent. Immunohistochemically, all cells and the muscle fibers showed strong positivity for desmin (Fig. 2a) including positivity of striated muscle cells (Fig. 2b) as well as for vimentin. Weak to moderate S100β immunoreaction was seen in about 40% of tumor cells (Fig. 2c) Reactions for cytokeratins (MNF116, CK7), GFAP, EMA, smooth muscle actin, and CD34 were negative in the tumor cells. Neurofilament positive nerve fibers were not seen. The Ki67/MIB-1 proliferation index was lower than 2%. **What is the diagnosis?**

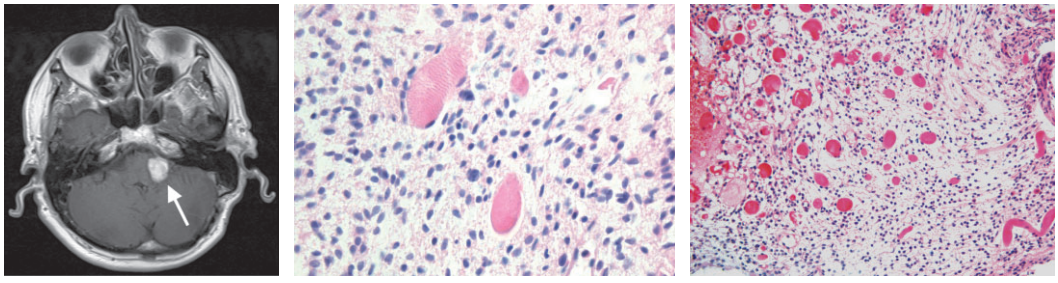

**Figure 1a,b,c.**

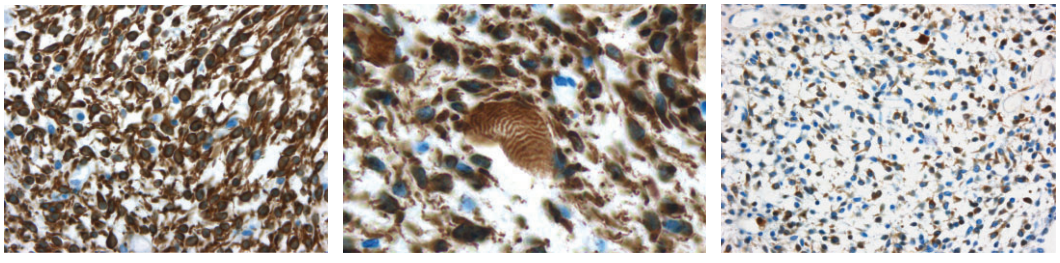

**Figure 2a,b,c.**

## DIAGNOSIS

Fetal type myxoid rhabdomyoma of the vestibular nerve.

## DISCUSSION

Rhabdomyomas are benign tumors containing striated muscle and are categorized by location as cardiac and extracardiac tumors. Cardiac rhabdomyomas typically occur in young patients and are often associated with tuberous sclerosis. Among extracardiac rhabdomyomas, adult, fetal and genital types are distinguished with head and neck region as the most common site. Whereas adult-type extracardiac rhabdomyomas mostly occur in adults older than 40 years, fetal-type tumors tend to occur predominantly in infants and may be associated with autosomal dominant nevoid basal cell carcinoma syndrome (Gorlin syndrome) characterized by *PTCH* mutations. Reports of rhabdomyomas manifesting at cranial nerves are exceptional, and only 4 cases have been published: Zwick *et al*. reported a rhabdomyoma of the left trigeminal nerve in a 29-month-old male infant replacing the 5th nerve in the posterior fossa and in the region of Meckel's cave. (7) Another report reviewed a case of a 41-year-old man with a tumor emerging from the right the porus acusticus internus, suggesting a schwannoma. (6) This tumor infiltrated the facial nerve root and extended from the brainstem to the geniculate ganglion. Histologically, many mature striated fibers were seen, and the tumor was localized within the nerve, which complicated the differential diagnosis of a schwannoma. However, Antoni A and B regions were not seen. A third case report described a rhabdomyoma of the left 8th cranial nerve in a 6-year old boy manifesting in the cerebellopontine angle with growth into the internal acoustic meatus and adhesion to the pons. (5) In a fourth case a boy presented with a rhabdomyoma of the left oculomotor nerve being attached to the midbrain. (2) These cases of intracranial nerve rhabdomyomas were referred to as adult type rhabdomyomas. In contrast, our case affecting a 68-year-old man represents a fetal type rhabdomyoma. The fetal type myxoid rhabdomyoma has been described mainly in infants and is especially localized subcutaneously in the preauricular and postauricular region thus indicating an exceptional localization. (1) Moreover, a fetal type rhabdomyoma occurring in adults seems unusual. Fetal type rhabdomyomas have variable histological patterns, with myxoid/classic and intermediate/cellular forms being distinguished. The myxoid/classic type mainly affects boys in the first year of life. Kapadia *et al*. reported 8 cases of myxoid/classic rhabdomyomas of which 5 cases had been younger than one year of age. (1) Yet, there was one adult female case of 20 years of age. Among all fetal rhabdomyomas of any type age ranged from 3 days up to 58 years. (1) Thus, fetal type myxoid rhabdomyomas can occur at later stage in life.

In principle, rhabdomyomas occurring in association with nerves, should be distinguished from neuromuscular hamartomas (choristomas), also called intraneural rhabdomyoma-like tumors. These lesions are non-neoplastic. They are composed of well differentiated mature skeletal muscle cells and peripheral nerve tissue and are discussed to arise from displaced muscle fibers during embryonal development or to originate from neuroectodermal cells. Neuromuscular hamartomas (choristomas) are synonymously called benign triton tumors in the literature although some authors have proposed that this term has been incorrectly applied. They suggest that benign triton tumors represent neurofibromas with rhabdomyomatous component. (3, 4)

Although histological criteria in general help to distinguish rhabdomyomas from neuromuscular hamartomas, it may be difficult in certain cases. In particular, clear differentiation between adult type rhabdomyomas associated with nerves and neuromuscular hamartomas may be challenging as reflected for 3 out of 4 reported cases of cranial nerve rhabdomyomas. (2, 6, 7) It may be important for the managements of patients to distinguish between hamartomas and true tumors, both occurring at cranial nerves. A literature review revealed 16 patients with neuromuscular hamartomas/choristomas of cranial nerves involving optic, oculomotor, trigeminal, vestibular, maxillary and facial nerves. Concerning our case, myxoid stroma, absence of nerve structures and nerve fibers as well as recurrence is in concordance with the diagnosis of a fetal type, myxoid rhabdomyoma and excludes a neuromuscular hamartoma or benign triton tumor. Mitoses were absent in our case as has been described previously. (2, 5)

Since only few cases of intracranial rhabdomyoma are reported the prognosis is not clear. Follow-up was reported in only one case with no recurrence after four years. (5) Nevertheless our case demonstrates that recurrence can occur due to incomplete resection.

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## ABSTRACT


We report on a 68-year-old male with a cerebellopontine angle tumor manifesting at the 8th cranial nerve and presenting histopathological features of a rhabdomyoma. A literature review revealed four reports of intracranial nerve rhabdomyoma, all of adult type and including one manifestation at the vestibular nerve. We present the first case of a fetal type extracardiac rhabdomyoma manifesting at a cranial nerve. Although rare, rhabdomyomas must be considered in the differential diagnosis of vestibular schwannomas.